GEORGIAN MEDICAL NEWS

ISSN 1512-0112

NO 3 (336) Март 2023

ТБИЛИСИ - NEW YORK

ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии

საქართველოს სამედიცინო სიახლენი

GEORGIAN MEDICAL NEWS
No 3 (336) 2023

DELAYED SURGERY FOR GIANT SPONTANEOUS RUPTURE OF THE DISTAL THORACIC AORTA CAUSED BY CYSTIC MEDIAL NECROSIS

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Abstract.

Spontaneous (idiopathic) thoracic aortic rupture (STAR) is uncommon and assumes a rupture of the normal-sized thoracic aorta with no visually apparent aortic disease. Since 1961 about fifty reports have been published. STAR is established in cases of thoracic normal-sized aorta rupture with no traumas, aneurysms / dissections, infection, inflammation, connective tissue diseases, aortic and adjacent organs tumors/metastases, previous surgery, and occurring during pregnancy and the peripartum. Atherosclerosis penetrated atherosclerotic ulcer, neurofibromatosis type I, peri- and postpartum estrogen-mediated elastin irregularities of the aortic media, and cystic medial necrosis (CMN) were identified as a cause of STAR when histopathological examinations were performed. A case of a 68-year-old man with giant STAR of the descending aorta in the background of CMN is reported here. The patient in terminal hemodynamic condition was successfully treated by delayed open surgery two weeks after the disease's onset.

Key words. Aortic rupture, spontaneous, thoracic aorta, acute aortic emergencies, Gsell-Erdheim's syndrome, cystic medial necrosis.

Introduction.

Spontaneous or idiopathic rupture of the thoracic aorta is a sudden rupture of the normal-sized aorta with no visually apparent aortic disease [1,2].

Spontaneous thoracic aortic rupture (STAR) is an uncommon event. Since 1961 up to 2006 only 31 cases have been reported [1]. It was found only 19 additional cases on Medline for the period between 2006 – 2023 (Table 1). Due to its high potential to be lethal, STAR demands to be treated urgently. Since only successful cases are usually published, it can be assumed that the real number of cases is an underestimate.

A case of a giant spontaneous rupture of the distal descending thoracic aorta, which occurred in the background of cystic medial necrosis is reported here. The patient in terminal hemodynamic condition was successfully treated by open surgery two weeks after the disease's onset.

Case report.

A 68-year-old man experienced an acute chest pain attack two weeks before admission to our hospital. In the small too remote hospital in the place where the patient lived, CT scans on the day of disease's onset revealed the normal-sized thoracic aorta with local lateral protrusion in the aortic contour of the distal segment of the descendent thoracic aorta (Figure 1A,1B and 1C). Thoracic aorta rupture was recognized and treated by immobilization, the maintenance of systolic blood pressure not above 100 mm Hg, transfusions of fresh frozen plasma / red cells and the drainage of the left chest cavity. There was no equipment and technical opportunities for endovascular or surgical procedural treatment in the hospital. The search for vascular centers and patient transportation took as long as two weeks. As a result, the patient was admitted to our hospital two weeks after disease onset in poor clinical condition with depressed consciousness, a weakened peripheral pulse, borderline hemodynamics, and limited urinary output. There was significant increasing of paraaortic hematoma and bilateral pleural effusion on the CT at the admission (Figure 1D). An endovascular approach was not considered due to the poor condition of the patient demanding circulatory support. Immediately, the left thoracotomy was performed via the 6th intercostal space with evacuation of 1500 ml bloody fluids. The descending thoracic aorta was surrounded by a thick layer of blood-imbibed paraaortic tissues. The aorta was cannulated just after the left subclavian artery branching. The main pulmonary artery was cannulated intrapericardially for venous return. The cardiopulmonary bypass (CPB) was initiated, and the patient's temperature decreased to19°C. The CPB volume was diminished to 0, 5 l/m2. The proximal aortic clamp was placed on the free aorta just distal to the cannulation site. The distal thoracic aorta was opened without clamping. The intima appeared non-atherosclerotic. The X-shaped rapture measured at 35 x 50 mm was identified on the lateral wall in accordance with the protrusion on the CT (Figure 2A). The ruptured segment was replaced with a 120mm-long linear prosthesis (Figure 2B). After aortic unclamping, the CPB was restored to 2.5L/m2. During re-warming, the heartbeat has been spontaneously restored. The aortic cross-clamping period was 53 minutes. The patient was weaned from CPB. The aortic specimen revealed changes coincident with cystic medial necrosis (CMN) (Figure 2C and 2D). The patient's recovery was prolonged due to acute renal failure without replacement therapy and gastroenteropathy demanded of entire parenteral nutrition during the two weeks after the procedure. At 8-months follow-up, the light weakness of legs persists, demanding the use of a walker to be insured.

Discussion.

The diagnosis of spontaneous or idiopathic thoracic aortic rupture assumes a sudden rupture of the normal-sized aorta with no apparent disease of the aortic wall and no external impacts [1]. The diagnosis is highly heterogeneous. In the published reports of open surgical treatment, the thoracic aortic ruptures were named as 'spontaneous' if there were no traumas, aneurysms /

Table 1. *Review of literature (2005 – April 2023): Spontaneous Rupture of the thoracic aorta.*

Author(Year)	Age/Sex	Comorbidity	Rupture site	Phatology	Operation	Outcome
Komanapalli CB(2006) [1]	40M	N/A	Distal arch	Fragmentation of elastica in media	Open surgery (GR)	Survived
Hirai S (2006) [17]	72M	N/A	Ascending	N/A	Open surgery (GR)	Survived
Gaspar M (2007) [5]	53M	Hypertension	Aortic arch	Atherosclerosis	Open surgery (GR)	Survived
Yoshida M (2008) [3]	61M	Takayasu's arteritis	Descending	N/A	Endovascular stent	Survived
Brizzio ME (2009) [4]	N/A	N/A	Ascending	N/A	Open surgery (GR)	Survived
Belov YuV (2011) [6]	67F	Diabetes Mellitus, type I	Descending	Penetrated atherosclerotic ulcer	Open surgery (PR)	Survived
Sasaki H (2012) [18]	75M	N/A	Aortic arch	N/A	Open surgery (GR)	Survived
Lu W (2012) [19]	51M	N/A	Descending-abdominal	N/A	Endovascular stent	Survived
Park YJ (2012) [11]	49M	Neurofibromatosis (NF-1)	Descending-abdominal	Fragmentation of elastica in media	Open surgery (GR)	Survived
Menon A (2013) [7]	17F	Postpartum	Descending	Fragmentation of elastica in media	Open surgery (GR)	Survived
Itonaga T (2014) [20]	67M	N/A	N/A	N/A	Endovascular stent	Survived
Akchurin RS (2015) [21]	67M	Hypertension	Distal arch	N/A	Endovascular stent	Survived
Yoshida C (2015) [13]	94F	N/A	Descending	N/A	Open surgery (N/A)	Survived
Furukawa T(2018) [9]	N/A	Postpartum	N/A	N/A	Endovascular stent	Survived
Tateishi A (2019) [12]	N/A	Peripartum, Neurofibromatosis (NF-1)	Ascending	N/A	Open surgery (N/A)	Survived
Guner EG (2020) [8]	22F	Peripartum	Ascending	Fragmentation of elastica in media	Open surgery (ARR)	Survived
Kasahara H(2021) [2]	76M	N/A	Aortic arch	N/A	Endovascular stent	Survived
Yokoyama T (2021) [10]	N/A	N/A	N/A	N/A	Endovascular stent	Survived
Yokoyama T (2021) [10]	N/A	N/A	N/A	N/A	Endovascular stent	Survived

M – male; F – female; N/A. – not available; GR – Graft replacement; PR – Patch repair; ARR - Aortic root repair.

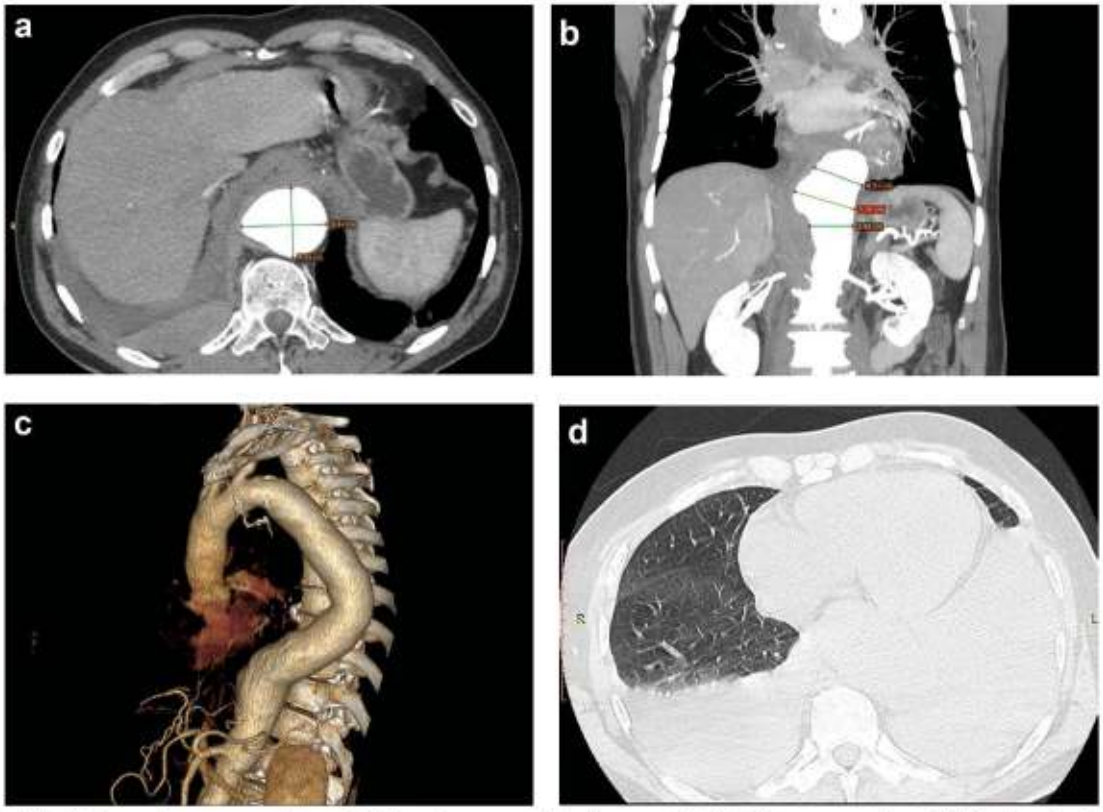

Figure 1. *(A) Axial, (B) Coronal contrast-enhanced CT at the onset of the disease; (C) 3D CT reconstruction; (D) Deferred axial CT scan after two weeks from disease onset.*

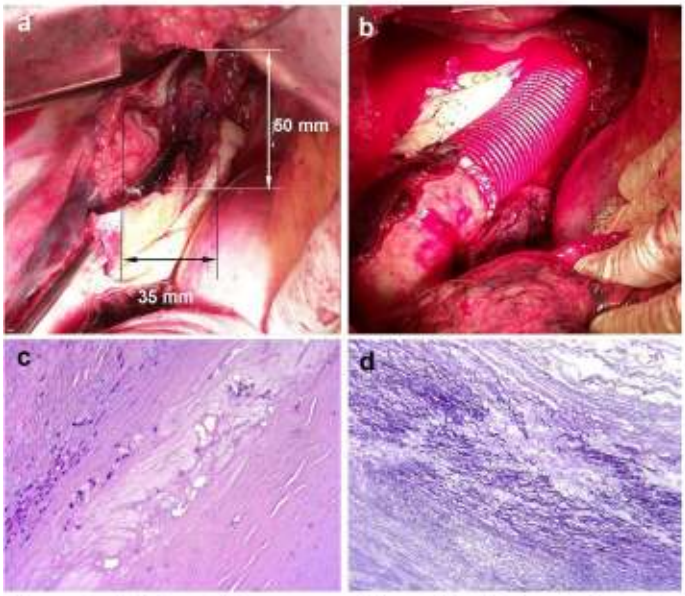

Figure 2. *(A) View of the rupture; (B) cyst-like lesions in the media, H&E×200; (C), elastic fibers disruptions, Weigert staining×100.*

dissections, infection, inflammation, connective tissue diseases, aortic and adjacent organs tumors/metastases [2,3], previous surgery [4] along a normal aortic size. Simultaneously, the penetrative atherosclerotic ulcer was indicated as a cause of aortic rupture also named as 'spontaneous' [5,6]. Aortic ruptures during pregnancy and the peripartum period because of progressive, estrogen-mediated elastin irregularities of the aortic media have also been reported as cases of STAR [7-9].

While endovascularly treated cases were reported with no histopathological examinations, the causes of the rupture were recognized in all surgically treated cases after specimen examination. Aortic wall abnormalities such as atherosclerosis [5,10], neurofibromatosis type I [11,12], and cystic medial necrosis were identified as a background in cases of STAR. There are no case reports on STAR with healthy aortic wall in histopathological examinations.

STAR is a rare event. Thirty-one cases treated surgically were founded to be reported from 1961 to 2004 [1,5]. The development of endovascular aortic repair has led to a growing number of reported cases of successful treatment within the last 15 years. Nineteen both surgically and endovascular cases have been discovered on Medline between 2005 and the present (Table 1). Since predominantly successful outcomes are usually published, the real number of cases may be underestimated.

Because of hemodynamic shock, prompt diagnosis and urgent intervention are necessary in the case of STAR [2]. CT is useful for prompt diagnosis, but it is sometimes difficult to detect the exact site of the lesion [2,10]. Any irregularity, ulcer-like projection, extravasation of contrast medium, prominence of aortic contour on CT may help to find the site of lesion. 3D reconstruction may also be useful [13] as in the case described here. The localization and extent of periaortic hematoma also helps to define the location of aortic rupture.

Treatment modalities include open surgical and endovascular techniques. Endovascular treatment has obvious advantages in comparison to open surgery due to the possibility of prompt use, less trauma, and no need for cardiopulmonary bypass. In conditions of extended rupture and absence of its exact localization, endovascular treatment demands a lengthened stent position with potential for spinal cord ischemia and associated paralysis of the lower extremities [10].

Because only singular or short serial cases with no long follow-up were published, it is not possible to describe STAR prognosis, so it is left obscured.

Cystic medial necrosis as well as initial lysell-Erdheim syndrome is a pathology of the aorta and great arteries characterized by degeneration of elastin fibers with loss of smooth muscle cells in the vascular medium that produces areas of glycosaminoglycans accumulation initially described as cysts. The term CMN is historic, since, in fact, no cysts or necrosis exist. The etiology is not well understood. There are two theories of CMN etiology: 1. CMN is a result of affecting on aortic wall of xenobiotic agents having excessive and growing environmental spread. 2. CMN is caused by high activity of tissue proteinases. The activity of tissue proteinases is regulated by numerous complex mechanisms including unrecognized. The result of both etiological pathways is degeneration of elastin fibers with loss of smooth muscle cells in the vascular medium with glycosaminoglycans accumulation [14]. This disease accounts for 0,09% of all sectional cases, occurs in 8.0–22.2% of patients with aortic dissection without Marfan syndrome and in 40–82.0% of patients with combined aortic dissection with Marfan syndrome [15]. The average age of patients ranges from 32 to 50 years, but cystic medionecrosis can be even in newborns and young children [15]. The correct prevalence of CMN is difficult to determine but is quite high as a background in connective tissue diseases such as Marfan syndrome, Ehlers-Danlos syndrome, annul aortic ectasia, and congenital heart defects [15]. In recent time, CMN is documented in unaffected individuals also [14].

The described case demonstrates that the strength of the mediastinal tissues restrained exsanguination in the pleural cavity for two weeks, making performing the delayed open surgery possible.

Open surgery demands the application of CPB. There is no distal aortic clamp in the case because of the vast paraaortic hematoma. Deep hypothermic perfusion was used to reduce blood flow. It may be supposed that the light weakness of the legs persisting for 8 months is caused by compression of the muscle innervation for a long time period in the borderline hemodynamic conditions.

Conclusion.

In conclusion, STAR is a sudden rupture of the normal-sized aorta with no apparent aortic disease. STAR occurs in patients with aortic wall background abnormalities. Only about fifty reports have been published in the literature up to the present. STAR is a life-threatening condition demanding urgent open or endovascular treatment. Giant aortic rupture here described has been presented with a long enough survival period due to the strength of the mediastinal tissues preventing exsanguination.

The delayed open surgery appeared successful despite poor initial clinical conditions of the patient.

The patient's consent was obtained to present the case and to provide images.

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